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Factitious Sickle Cell Acute Painful Episodes: A Secondary Type of Munchausen Syndrome

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Three adult patients with factitious acute sickle cell painful episodes are presented and the literature is reviewed. The prevalence of this disorder among patients with sickle cell disease in our program was found to be about 0.9%. The patients described to date were all young adults who demonstrated pathological lying (pseudologico fantastico) and most of them had an underlying authentic medical illness to which the feigned signs and symptoms of sickle cell disease were added, thus making the diagnosis more plausible. It is recommended that all patients who present themselves with the signs and symptoms of sickle cell painful episodes be carefully studied in order to confirm the diagnosis.

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INTRODUCTION

Individuals with factitious disorders present themselves with physical or psychological symptoms that are feigned or produced intentionally in order to assume the sick role [1]. The fourth edition of the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-IV) lists three criteria for the diagnosis of factitious disorders as follows [2]: (1) a sine qua non feature pertains to the *intentional* production of physical and/or psychological signs or symptoms. These may include fabricated subjective complaints, self-inflicted conditions, exacerbation or exaggeration of preexisting general medical conditions, or any combination thereof; (2) the major motivation for the factitious behavior is to assume the sick role; and (3) the absence of external incentives for the behavior.

Munchausen syndrome is an example of a factitious disorder with predominantly physical signs and symptoms [1]. An essential feature of this syndrome is a tendency for individuals to give a fabricated but plausible history and seek out care by wandering from hospital to hospital. Two patients who fabricated the signs and symptoms of sickle cell anemia on numerous occasions but who had normal hemoglobin electrophoresis were described by Lindenbaum [2]. He first used the terms hemoglobin Munchausen to designate this condition. A third example of hemoglobin Munchausen described a young man with hemolytic anemia and painful episodes ascribed to "sickle" but with normal hemoglobin electrophoresis [3]. A fourth example of Munchausen sickle cell painful crisis was described in a patient who repeatedly fabricated the signs and symptoms of acute painful episodes but was found to have sickle cell trait plus severe iron deficiency anemia that required blood transfusion [4].

The purpose of this report is to describe the experience of our sickle cell center with factitious sickle cell painful episodes over the last 20 years. The aim is to determine their prevalence, characteristics, and possible psychopathologic processes that predispose certain individuals to the intentional fabrication of the signs and symptoms of recurrent attacks of sickle cell pain.

MATERIALS AND METHODS

Patient Population

Between 1975 and 1995 we have seen a total of 424 adult patients with various types of sickle cell disorders in our center. Of these 287 had sickle cell anemia, 87 had Hb SC disease, 47 had sickle-β-thalassemia, and 3 had other combinations (Hb SD and Hb SO Arab). During

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the same period we saw four patients with factitious sickle cell painful episodes. One of these patients was previously reported [4] and the other three are described below.

CASE REPORT 1

A 32-year-old African American male was admitted to our hospital for the first time via the emergency room with the diagnosis of acute sickle cell painful episode involving his low back, right hip, and right ankle. He reported that the pain was similar to his previous crisis pain and had a categorical scale of 8/10. He stated that he had recently moved to Philadelphia from New York and felt that the stress of that situation may have precipitated the present painful event.

He gave past history of Hb SC disease that was diagnosed between the ages of 8 and 9 years when he became sick and had to be hospitalized. He related that he was usually hospitalized once every 8 weeks for painful crises. Each crisis lasted 7–14 days. The pain usually involved his low back, hips, knees, ankles, and wrists. The pain used to be treated with meperidine but recently he was switched to hydromorphone. He reported certain complications of his disease including cholelithiases, leg ulcer over the right ankle, history of splenectomy (the reason was not clear), history of retinopathy in both eyes, and S/P right hip replacement for avascular necrosis. Later he had osteomyelitis of the right hip and the prosthesis had to be removed. He gave no history of blood transfusion.

In addition to Hb SC disease he gave history of insulin dependent diabetes mellitus (which may have caused the retinopathy) and mild hypertension. These were managed with 35 U of NPH insulin subcutaneously (s.c.) every morning, 5 U NPH insulin s.c. in the afternoon, and nifedipine 30 mg extended release by mouth daily. Noteworthy is that during the interview there was a tendency to emphasize sickle cell disease over diabetes and hypertension. He indicated that his 5-year-old daughter also had Hb SC disease and that some of his friends in New York also had sickle cell disease.

Physical exam showed a healthy and cooperative young male in no distress. He walked with a limp to the right and used a walking cane. Vital signs were within normal, temperature was 36.8°C, systolic blood pressure was 136 mmHg and the diastolic 92 mmHg. Pertinent physical findings included absence of scleral icterus, no heart murmurs, clear lungs, a liver span of 10 cm, healed operative scars in the left upper quadrant, the left paraumbilical area, over the lateral aspect of the right hip and thigh, and a scar of healed ulcer over the right ankle.

Pertinent laboratory data and features of his disease are listed in Table I. Sequential multiple analyses of his serum (SMA-7 and 12) showed normal values of Na, K, Cl, CO_2, total and direct bilrubin, BUN, and creatinine. Glucose was mildly increased to 121 mg/dl, lactate dehydrogenase (LDH) 274 IU/L (N = 110–220), alkaline phosphatase 178 IU (N = 35–115), alanine aminotransferase (ALT) 47 IU/L (N = 1–45), asparate aminotransferase (AST) 67 IU/L (N = 1–36), and γ-glutamyltranspeptidase (GGT) 77 IU/L (N = 10–50). Urinalysis showed 3+ proteinuria and 24 hr urine protein excretion was 910 mg (N < 150). Radiographs of the right hip and pelvis showed absence of the femoral head and status post girdlestone procedure.

Noteworthy is that examination of the peripheral smear showed normal RBC morphology with *no* sickle cells, target cells, spherocytes, or "boat"-shaped cells that are typical of Hb SC disease. Hemoglobin electrophoresis was normal and no Hb S or C was detected in his blood.

Because his history was so plausible, it was decided to give him the benefit of the doubt and repeat the hemoglobin electrophoresis. His pain was managed with hydromorphone and he was discharged after 12 days of hospitalization. Follow-up as an outpatient again showed normal hemoglobin electrophoresis. At that point he was told that he did not have Hb SC disease and that his major medical problem was diabetes and mild hypertension. He left the office and was never seen again in our facility.

CASE REPORT 2

A 28-year-old African American female was admitted to our hospital for the first time via the emergency room with the diagnosis of acute sickle cell painful episode involving her low back and legs. She reported that the pain was similar to her previous crisis pain. She also gave history of a seizure disorder and complained of having had two seizures on the day of admission and two seizures on each of the two days preceding her admission. She stated that she had been instructed previously to take phenytoin and phenobarbital. However, she had not taken these medications for the last several weeks. She also indicated that she was 3 months pregnant.

She gave past history of sickle cell anemia that was first diagnosed in early childhood. She experienced about one painful episode per month that required treatment in the emergency room and/or hospital. The pain usually involved her back and legs and lasted up to 2 weeks. The crisis pain was usually treated with meperidine in the emergency room/hospital and with oxycodone plus codeine at home. She also had pneumonia in childhood, recurrent urinary tract infections, one episode of pyelonephritis, and cholelithiasis. She was transfused once in childhood. She indicated that her mother had sickle cell disease, that her father had sickle cell trait, and that her cousin had sickle cell anemia. The cousin in question did have sickle cell anemia and was enrolled in our sickle cell program. She denied the use of alcohol and admitted to previous use of crack and cocaine but denied any current use.

TABLE I. Clinical and Laboratory Characteristics of the Patients Studied*

	Patient 1	Patient 2	Patient 3
Age (Y)/sex	31 M	28 F	21 M
Hb (g/dl)	11.0	14.9	16.3
Hct (%)	34.4	40.9	49.1
MCV (fl)	85	85	94
Reticulocytes (%)	0.9	0.5	ND
WBC ($10^3/\mu l$)	4.9	6.0	9.3
Platelets ($10^3/\mu l$)	276	323	397
Hb S (%)	0	0	0
Hb C (%)	0	0	0
Hb F (%)	<2.0	<2.0	<2.0
Hb A_2 (%)	2.5	3.4	2.4
Hb A (%)	97.5	96.6	97.6
Coexistent diagnoses	IDDM Hypertension Healed leg Ulcer	Seizure disorder	Asthma Bone infarcts Secondary to steroids

*Hb, hemoglobin; Hct, hematocrit; MCV, mean corpuscular volume; WBC, white blood cell count; IDDM, insulin dependent diabetes mellitus.

Physical exam was noncontributory. Pertinent lab data are shown in Table I. Sequential multiple analysis of serum (SMA-7 and SMA-12) were all within normal limits. Urine and serum were positive for cocaine. Urine and serum pregnancy tests were negative. Examination of the cerebrospinal fluid showed no abnormality. Computed tomography of the head showed no abnormalities. EEG was abnormal due to the presence of non-specific generalized and paroxysmal slow activity with a left-sided predominance. No epileptiform abnormalities, however, were identified.

Management consisted of treating the seizure disorder with phenytoin and phenobarbital and her pain with meperidine based on the history. Attempts to change meperidine to other opioids in view of the history of seizure were refused by the patient.

Examination of the peripheral smear showed mild aniso- and poikilocytosis but no sickle cells were seen. Hemoglobin electrophoresis was normal with no Hb S or C detected.

At this point she was told that she did not have sickle cell anemia but that her seizure disorder needed further observation and work up. She did not receive this information well and left the hospital 7 days after admission and never came back to our facility.

CASE REPORT 3

A 21-year-old African American male was admitted to our hospital for the first time via the emergency room with the diagnosis of acute sickle cell painful episode involving his low back, legs, and joints. He was vague about details of his sickle cell disease and its complications but indicated that he had avascular necrosis of his right hip. He also stated that his father had sickle cell disease and that his mother had sickle cell trait. Noteworthy was that his significant other was known to have Hb SC disease and was enrolled in our sickle cell program. As an outpatient he took meperidine 50 mg orally as needed.

Past history was significant for asthma since childhood. Treatment of asthma included bronchodilators and prednisone 80 mg by mouth daily. He mentioned that he had to be intubated once in the past and that he had peptic ulcer disease due to steroids.

Physical exam was non-contributory. Pertinent lab data are shown in Table I. Skeletal X-rays showed mild degenerative changes in the mid cervical spine and bilateral proximal and distal tibial metaphyseal infarcts with no definite evidence of avascular necrosis of the femoral heads. These changes may have been the result of chronic therapy with high doses of prednisone.

Examination of the peripheral smear showed no sickle cells and the red cell morphology was within normal. Hb electrophoresis showed no Hb S or Hb C (Table I). Communication with his family physician confirmed the history of asthma and its treatment and revealed that he was known to have factitious febrile syndrome and that he consumed relatively large amounts of meperidine for his "bone pain."

DISCUSSION

Munchausen Syndrome is a term first introduced by Asher [5] for Baron Hieronymus Karl Friedrich von Munchausen, a storyteller of alleged heroic deeds in the German cavalry. Patients with this syndrome deceive health care providers by simulating signs and symptoms of physical illness for no obvious gain. In some cases, similar

TABLE II. Characteristics of Reported Patients With Factitious Sickle Cell Disease*

Patient	Age (Y)	Sex	Feigned diagnosis	Coexistent disease	Exposure to patients with SCD	Pathological lying	Reference
1	31	M	Hb SC	IDDM Hypertension	Yes; friends	Yes	This report
2	28	F	SS	Seizure disorder	Yes; cousin	Yes	This report
3	21	M	SS	Asthma Bone infarcts	Yes; significant other	Yes	This report
4	22	F	SS	Severe IDA (Hb AS)	Yes; nephew and cousin	Yes	[4]
5	28	F	SS	IDA, UTI (Hb AC)	Yes; she is a nurses' aide	Yes	[2]
6	22	F	Hb SC	PID	Probably yes; other patients	Yes	[2]
7	20	M	"Sickle"	Hemolytic anemia	Probably yes; other patients	Yes	[3]

*Hb SC, hemoglobin SC disease; SS, sickle cell anemia; Hb AS, sickle cell trait; Hb AC, hemoglobin C trait; IDDM, insulin dependent diabetes mellitus; IDA, iron deficiency anemia; UTI, urinary tract infection; PID, pelvic inflammatory disease; SCD, sickle cell disease.

to the first patient described in this study, the clinical picture could be so plausible that patients might receive treatment for the alleged disorder on a chronic basis. It is quite understandable how a patient like this would smoothly fit into a care program on a continuous basis if no hemoglobin electrophoresis is performed. Munchausen patients often exhibit surgical scars from unnecessary operations by previously deceived physicians. This may or may not have been the case in the first patient described in this study as past history was unavailable to verify the necessity of the "alleged" splenectomy and hip replacement. Because our program requires establishing the exact type of sickle cell disease (i.e., sickle cell anemia, Hb SC disease, sickle-β-thalassemia), we perform hemoglobin electrophoresis on all patients who present themselves with this diagnosis.

Munchausen Syndrome has been divided into two subtypes [6]: prototypical and non-prototypical. Patients with the first type are usually unemployed males who drift from hospital to hospital. Patients with the non-prototypical syndrome could be either males or females who often simulate illness and who often stay in one place for months or years. The patients described here seem to fit into the non-prototypical type of Munchausen Syndrome. It should be noted that Munchausen Syndrome is distinct from malingering. In the latter disorder the motivation for the symptom production is secondary gain which is usually an obvious external incentive. Malingerers, for example, may seek hospitalization by producing symptoms in an attempt to get "room and board" or obtain compensation and they can discontinue this behavior when the symptoms are no longer useful to them [1].

Between 1975 and 1995 we saw 424 adult patients with sickle cell disease in our center. During the same period we saw four additional patients with factitious sickle cell painful episodes: 3 in this study and one previously reported [4]. Thus the prevalence rate of Munchausen Syndrome among our adult patients with sickle cell disease is about 0.9%. Whether other sickle cell centers have a similar prevalence rate of Munchausen Syndrome among their patients or not is unknown at the present time. Munchausen patients are known to visit a number of different health care facilities at various geographic locations over many years. It is likely, therefore, that the actual prevalence rate of pseudo-sickle cell patients in the United States is smaller than estimated in this study.

The characteristics of the seven patients with factitious sickle cell disease are summarized in Table II. All the patients are young adults who demonstrated pathological lying (pseudologica fantastica). It is interesting to note that most of the patients had other authentic disease processes but the patients choose to transpose their diagnosis to sickle cell disease. This is unlike the classical description of Munchausen Syndrome where the patients are basically disease free and they feign symptoms in order to obtain treatment. In Munchausen sickle cell disease, on the other hand, most of the patients seem to have an authentic disease process but they elect to change their diagnosis into painful episodes and feign the appropriate syndromes thus making their history plausible in order to obtain the desired treatment. Perhaps this type of disease process should be called Secondary Munchausen Syndrome in contradistinction to the primary type where there is no underlying disease process of any type.

The motives for Munchausen Syndrome are complex and a specific motive in a certain patient may never be clearly defined [5]. The primary motive in the patients with factitious sickle cell crises described most likely includes the desire to be the center of attention. Other motives such as the desire to obtain free room and board [7] and obtain opioid analgesics are unlikely secondary incentives. It is also conceivable that some of the patients were truly convinced that they had sickle cell disease because they were misinformed repeatedly by previous physicians who might have diagnosed them without the benefit of hemoglobin electropheresis. Individuals with sickle cell trait are known to develop symptoms when

they misunderstand the diagnosis or misinterpret the information given in counseling.

Finally, it is to be emphasized that patients who present themselves with signs and symptoms of sickle cell disease should be properly tested in order to confirm the diagnosis. This confirmation becomes particularly important prior to major interventions such as surgery and prior to the repeated use of opioid analgesics.

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